HEMORRHAGIC INTERNAL PACHYMENINGITIS IN CHILDREN.

REPORT OF TWO CASES.

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Most writers upon nervous diseases who make any mention of the occurrence of chronic internal pachymeningitis in children regard it as a rare occurrence. Sachs,[1] in his recent text-book, does not describe the condition, and I have known the existence of the lesion in children to be questioned by experienced neurologists. On the other hand, Doehle,[2] of Kiel, found a new membrane attached to the inner surface of the dura in 48 out of 597 autopsies on children under ten years of age—that is, in more than 17 per cent. of his autopsies. Although these figures are surprisingly high, it seems probable that chronic internal pachymeningitis is hardly to be regarded as a rarity in hospitals for children, and that the lesion in its slighter grades may be readily overlooked. Certain it is that the lesion is sufficiently frequent to deserve more attention than it has received. It is with a view to increasing the interest in this condition, which is not without practical importance, that I desire to report the following cases of chronic internal pachymeningitis, each of which was characterized by the occurrence of hemorrhage.

Case I.—A female child, aged five and one-half months, was admitted to Dr. Kimball's service in the Babies' Hospital on May 15, 1897. The family history was entirely negative. The patient was the first child. It is said that the child was well until one month ago, when it began to vomit, and vomited frequently until a short time previous to admission. The bowels were constipated. Examination showed that the child was fat and well nourished. The head was about normal in size and shape. The fontanelle measured two by two inches, and was not bulging. The sutures were open, and there was a soft spot over one parietal bone. The eyes, ears, nose, and mouth were normal. There were no teeth. Coarse râles were heard throughout the chest. The child vomited on the third and fourth days after admission. On the fifth day she developed tremor in the hands, especially marked in the right hand. The fontanelle was at this time rather tense, and the veins of the scalp distended. From time to time nystagmus was present, lateral and vertical in character. The pupils were small and responded feebly to light at times.

Nine days after admission the child had a general convulsion, which began with twitching in the ocular muscles and extended to the hands and feet. During the seizure the left arm and leg were rigid, while the right arm and leg were the seat of clonic spasm. The mouth deviated to the left. The duration of the seizure was about ten minutes. Cyanosis

[1] Nervous Diseases of Children, 1895.

[2] Verh. d. X. Internat. Congress, 1890, Bd. v. (Forensic Section).

was a feature of the convulsion. Ten hours after the first convulsion a second occurred, lasting about three minutes and closely resembling the first. After these seizures it was noted that the fontanelle was slightly sunken, and that the head constantly deviated to the right. Previous to these paroxysms the child had been drowsy; it now became semi-comatose for a time, but later was wakeful. The child grew gradually weaker, owing partly to the persistence of a severe colitis, which was the cause of the admission to the hospital, and after a few days died without having developed any new symptoms of neurological interest. The pulse and respiration were featureless and such as one might expect in any young child whose temperature ranged irregularly from 100° to 105°. During the last few days there were present the usual signs of bronchitis.

Autopsy by Dr. Woolstein, thirty-three hours post mortem.

Anatomical Diagnosis. Hemorrhagic pachymeningitis; fibrinopurulent pleurisy; acute broncho-pneumonia; pulmonary emphysema and congestion; fatty liver; nephritis.

Body. Plump, no skin lesions. Thumbs adducted, fingers loosely flexed.

Brain. Adhesions between pia and dura along superior longitudinal fissure, also over entire base and over the island of Reil on both sides. Covering the pia at these points there is a membrane, two to three lines in thickness, containing many spirally coiled bloodvessels, all filled with blood.

Pia apparently not adherent to brain. Ventricles normal in size; they contain about one drachm of fluid, hemorrhagic in color.

Choroid plexuses intensely congested, fluid blood in all the sinuses. Pia over cerebellum, pons, and medulla slightly thickened.

Cervical cord shows same conditions; rest of cord not examined.

Cerebrum. Right occipital region; pia detached from cortex in many places, here and there pia stained brown by pigment from extravasated blood. There is perceptible in this region, though more marked in sections from the island of Reil, a splitting up of the membrane overlying the cortex into two or more layers; the inner layer (pia) shows infiltration with small round cells, the layers external to this contain many new bloodvessels, some of which having ruptured, extravasation of blood into the surrounding tissues has occurred. Beside numerous small bloodvessels, the outer membranous layers consist of small round cells, fibroblasts, and a few very fine connective-tissue fibres. Between the layers lies a fine loose meshwork of small round and spindle cells containing a considerable amount of granular material (degenerated red blood-cells and fibrin).

Island of Reil. Same conditions, but much more marked.

Cerebellum. As above; formation of new bloodvessels very distinctly shown.

Choroid Plexus. Granular degeneration of epithelium and marked congestion.

Spinal Cord. Only slight traces of hemorrhage; membrane covering cord shows fibrous thickening; in places there is a division into two layers.

Case II.—Female infant, colored, aged twenty-two months, illegitimate, first child; mother healthy. Nursed for seven months. Since nursing was stopped has had repeated attacks of bronchitis. The child never walked or stood alone.

Examination: The head is markedly rhachitic, square, and flat on top. The anterior fontanelle measures three by two inches. The ribs are beaded; there is a marked lateral depression of the chest, and the epiphyses are enlarged.

The child's first stay in the hospital, about two months in duration, was featureless except for the facts that there were gradual slight loss in weight, considerable prostration, and numerous râles over both sides of the chest. During a second stay in the hospital, lasting a month, the child gained weight and showed no signs of bronchitis.

In October, 1897, the patient was admitted for a third time to Dr. Holt's service. It is stated that she was well until three days before admission, when she had four convulsions, followed the next day by five similar seizures. The general condition was very bad. There were signs of general bronchitis. The hands and feet were in the position of persistent flexor contraction characteristic of tetany. The knee-muscles were slightly rigid. The knee-jerks were unobtainable, and the fontanelle was tense and bulging. The child was in the hospital for five days before death occurred. During this time it grew very drowsy, but this was attributed to the administration of bromides. Hot baths had no effect on the carpo-pedal spasm. There was slight but varying rigidity of the muscles of the back of the neck. Ocular symptoms were not observed. During the last days of life the rigidity of the hands grew less, although there was no alteration in the spasm of the feet and legs. The child died comatose. Bloody mucous diarrhœa was present during the last period in the hospital. Fever was present and daily reached 102.5° or 103.5°.

Autopsy by Dr. Woolstein.

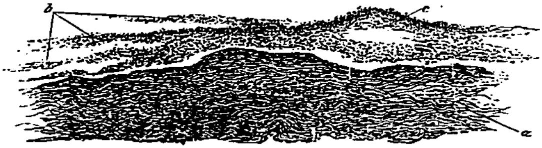
ANATOMICAL DIAGNOSIS. Pachymeningitis hæmorrhagica interna; broncho-pneumonia (acute and chronic); croupous and ulcerative colitis.

Brain. Weight, 1 pound, 5½ ounces (fontanelle 1½ x 1 inch; bulging). Over right half of convexity, a recent clot, in places one-quarter inch in thickness, covers the entire hemisphere, extending also on its outer surface and over the left occipital lobe. Upon being removed the inner surface of the dura is found lined by a thin, whitish translucent membrane, extending irregularly from the superior longitudinal fissure on either side. Dura thickened and yellow in color. The clot dips between the hemispheres and appears above optic commissure on the base.

Pia congested, and in places moderately œdematous. Ventricles and brain substance apparently normal. All sinuses filled with recent clots. The new membrane is especially distinct in both occipital fossæ.

MICROSCOPIC EXAMINATION. *Cerebrum* (right third frontal convolution and right temporo-sphenoidal lobe). There is some thickening of the pia, due partly to œdema, partly to infiltration with small round cells. The vessels of the pia are congested. Overlying the pia and connected with it by a fine web of branching cells and fibres is a membrane which in places shows distinctly a division into two or three layers. This membrane consists of round and spindle-shaped cells, with here and there a few very fine fibres; numerous small bloodvessels are seen, and hemorrhages have occurred into the meshes of the membrane, and caught within these meshes are red blood-cells, granular fibrin, and brown pigment. In places there are aggregations of small round cells, whose nuclei, staining very deeply with hæmatoxylin, are undergoing fragmentation (pus); these cells are found chiefly in the superficial layers of the membrane. At points where there has been considerable extravasation of blood the large branching connective-tissue cells are seen to be loaded with brown pigment.

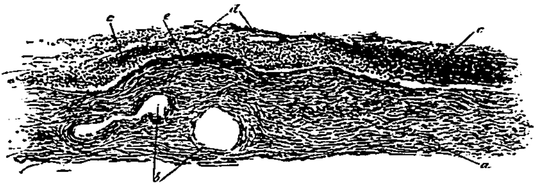
FIG. 1.



a. Dura. b. Layers of membrane. c. Purulent infiltration. Section through dura and new membrane in Case II.

Dura (near superior longitudinal sinus). The fibres are separated from each other by serous infiltration. The dura is covered by a membrane which in some places is thick, in others shows a subdivision into several layers. The membrane resembles that above described. Penetrating between the fibre bundles of the dura are seen fine strands of young connective tissue. Sections of dura taken from other regions show very extensive hemorrhages and brown pigmentation.

FIG. 2.



a. Dura. b. Bloodvessels. c. Hemorrhage into meshes of membrane. d. New vessels. e. Pigment. Section through dura and new membrane in Case II.

These cases illustrate some points in the etiology, clinical history, and pathology of chronic internal pachymeningitis which seem worthy of consideration.

The age of the first patient, five and one-half months, corresponds very nearly to the period of life at which this lesion is most frequent, for of 57 cases in which the age is given, 43 occurred under one year, and of these 34 were not more than six months of age. In a case reported by Northrup,[1] the patient was four years and seven months old, and in one mentioned by Doehle six years, and in another three and one-half years. These are the oldest children of whom I have found records. The condition is thus one relating to early infancy.

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[1] Transactions of the New York Pathological Society.

The majority of infants in whom the lesion is found are badly nourished or cachectic. Rickets was present in at least 13 of 57 cases, and chronic intestinal catarrh in at least 20 of these cases. In eight of Doehle's cases indications of syphilis were present. In the remainder of the 57 cases there is no record of syphilitic lesions or of syphilitic heredity, but in many of these such heredity cannot be excluded. In a few cases the children have been well nourished and apparently normal a short time before the indications of hemorrhage have appeared. As regards the pathological nature of the lesion we are as much in the dark as in the case of the similar lesion with which we meet in adults. The new membrane must be regarded as originating from the proliferation of dural endothelial cells, or, more probably, from the subendothelial connective-tissue cells, the new and cellular connective tissue being exceedingly prone to the development of new bloodvessels with thin walls. Even in cases which we may suppose to originate from hemorrhage we have to recognize an intimate connection between the organization of the clot and the proliferation of dural connective-tissue cells. In some cases there seems to be little inclination to hemorrhage, in others there are numerous punctate hemorrhages from the delicate vascular membrane which constitutes the inner layer of the laminated structure, and occasionally there is an extensive clot which lies in the subdural space, as in the second case reported. In the first case reported the membrane was extremely vascular, and small hemorrhages occurred into the membrane itself; in the second case the vascularity of the membrane was relatively slight.

The membrane varies much in thickness. In many cases it is so thin and delicate that it is readily overlooked unless the occurrence of the lesion be kept in mind. Sometimes it reaches a thickness of two or three lines. In both the cases reported here the membrane was extensive. In both cases it extended from the calvarium to the basal fossæ, and in one it was especially distinct in the occipital fossæ. Doehle noted that the membrane was especially apt to occur in the basal fossæ. He is strongly inclined to look upon the formation of the membrane as secondary to a meningeal hemorrhage at the time of birth, but the evidence for this view cannot be said to be conclusive. The belief that the lesions of internal pachymeningitis may be the consequence of subdural hemorrhage has recently received some support from Wigglesworth,[1] who pointed out the likelihood of hemorrhage arising from degenerated arachnoid veins in early life. It is stated that the experimental introduction of blood into the subdural space has been followed by the lesions of pachymeningitis. Although we are in no position to deny that extravasated blood may sometimes occasion the formation of the dural membrane, we cannot reasonably look on this as the explanation of the majority of the cases observed in infancy, in which there have apparently been no clinical signs of meningeal hemorrhage. In our first case, for example, the child was apparently normal until a short time before cerebral symptoms appeared, and the supposition of the existence of a meningeal hemorrhage at birth is hardly tenable. In Case II. considerable pigment was found near the line of attachment of the membrane to the dura, some free, some in the bodies of branching connective-tissue cells. It is not possible to interpret the relation of this pigment to the large hemorrhage, but there is no good reason for thinking that the locality of this pigment, which appears recent, indicates that the membrane originated from a layer of blood on the inner surface of the dura. The old experiments of Kremiansky,[1] who succeeded in producing some of the lesions of convexity internal pachymeningitis in dogs, by means of chronic alcoholic intoxication, indicates that an intoxication may give rise to the formation of a highly vascular membrane consisting of young connective-tissue cells and vessels. In many of the cases recorded in literature there is reason to suspect that pathological blood states may have existed, and that intoxications may have played a rôle in the development of the intradural lesion. But it is so common to find severe intoxications without such lesions that this does not seem a wholly satisfactory explanation. The intoxication theory also fails to explain the occurrence of the membrane in apparently healthy children. It may be that future pathological and clinical observations, together with experimental studies looking especially to the nature of the irritants which are competent to incite the dural connective tissue to proliferation, will show that the lesions with which we meet may be sometimes of inflammatory, sometimes of hemorrhagic origin, or, to be more accurate, will show that hemorrhage is one of the causes of the inflammatory lesion which we call chronic internal pachymeningitis.

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[1] Brain, 1892.

In sections from Case II., in which the membrane is less richly cellular and vascular than in Case I., the inner or free edge of the membrane is infiltrated with small spheroidal cells, whose cell-bodies are indistinct and whose nuclei are multiple and usually rounded and very distinct. The cells are probably leucocytes in which nuclear fragmentation has occurred. We are almost justified in speaking of the inner layers of the membrane as being the seat of a purulent inflammation, although the surface of the membrane was not covered with pus. I have met with no description of similar appearances in pachymeningitis, and the coincidence of such an exudative inflammation with the usual lesions of hemorrhagic pachymeningitis is certainly of much pathological interest. Doehle expresses the belief that the condition of marasmus seen in many of his cases may have been due to a trophic disturbance of the cortex cells dependent on damage by the pachymeningitis. In view of the frequency of intestinal catarrh in these patients Doehle's explanation seems superfluous; but it would, nevertheless, be interesting to look carefully for indications of structural change in the cortex which might be attributable to the influence of the dural lesion.

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[1] Virchow's Archiv, Bl. 68.

The presence even of an extensive and moderately thick membrane seems not to entail symptoms. If it does we have yet to learn what they are. It is probably impossible to recognize the condition until hemorrhage occurs, and even then the diagnosis is rarely to be made with confidence. The reported cases are not sufficiently numerous to enable us to study statistically the symptoms that accompany hemorrhage, but an idea may be formed of the character of the usual clinical phenomena. Where small hemorrhages from the newly formed vessels occur into the membrane itself there are probably no obtrusive cerebral symptoms. In a number of such cases the occurrence of a cerebral lesion has not been suspected during life. It is quite likely that slight cerebral symptoms in such cases are masked by the habitual apathy of these very young and usually marantic children. Where a hemorrhage of considerable size occurs, either into the new membrane or chiefly into the subdural space, the symptoms of irritation or compression of the cerebral cortex appear to be seldom absent. The hemorrhage is probably more often one-sided than bilateral, and the symptoms are generally correspondingly unilateral. The most frequent symptoms, where the hemorrhage is large, are rigidity, convulsions, and coma. If the hemorrhage occurs slowly convulsions may be absent, and rigidity with deepening coma constitute the only symptoms. When convulsions occur they may be repeated in groups a few days apart, sometimes owing to the occurrence of repeated hemorrhages of moderate size. Paralysis was noted in only a small portion of the hemorrhagic cases. Contracted pupils were observed in three out of nine cases. Tremor in the side opposite the lesion was seen in the first case. The symptoms of tetany were observed in the second case, and certainly constitute a feature of considerable interest. It is true that the phenomena of Trousseau and of Chvostek were not sought for, but the carpo-pedal spasm was so persistent that so careful an observer as Dr. Holt looked upon the condition as tetany. Carpo-pedal spasm or tetany is not rare without hemorrhagic pachymeningitis, and it does not seem certain that the spasm is in this instance to be referred to the lesion.

The fontanelle is usually bulging at the time of the onset of cerebral symptoms. The temperature is moderately elevated, the pyrexia being probably less than in the case of meningitis. Febrile complications, however, are so common that the temperature, pulse, and respiration of uncomplicated cases can rarely be studied. In fatal cases the duration of the cerebral symptoms is from a day to a week.

It is evident from the foregoing description of the symptoms of hemorrhagic internal pachymeningitis, that we can hardly hope to make the diagnosis with any degree of certainty. An extensive pial hemorrhage occurring in the course of an acute infectious disease, such as pneumonia, may apparently cause closely similar phenomena. I do not feel certain that there is any symptom or combination of symptoms which we meet in hemorrhagic internal pachymeningitis which we may not occasionally encounter in the course of a severe acute infection without the presence of any cerebral lesion whatever. At the same time hemorrhagic pachymeningitis is a lesion which we should think of as a possibility whenever we meet with unilateral rigidity and convulsions, with deepening stupor, in a cachectic or rhachitic child under one year of age. In the absence of the signs of pneumonia and tuberculosis, I think this diagnosis gains distinctly in probability, as we may then exclude with some confidence an important cause of infantile meningeal hemorrhage and also tubercular intracranial lesions.

Holt[1] has called attention to hemorrhagic pachymeningitis as a cause of the chronic cerebral palsies of children. I do not think it has yet been proven that this is one of the ways in which these palsies arise; but if it be true that children occasionally survive a hemorrhage from pachymeningitis sufficiently long to produce a chronic palsy, the anatomical evidence of this will surely be forthcoming.

One other aspect of internal pachymeningitis in children deserves mention. It is probable that relatively slight traumatisms to the head may occasion the rupture of vessels in a highly vascular membrane. This fact gives these cases a certain medico-legal importance. Doehle reports the case of a careless mother who allowed her apparently healthy infant to fall out of bed. The child was found comatose and died. The autopsy showed that the child had a hemorrhage from a subdural membrane. The question of the influence of trauma may thus arise.

Although it cannot be contended that hemorrhagic pachymeningitis in children is a frequent condition, even in hospitals for children, there are no doubt more examples of it than is generally supposed, and it is likely that its precursor, chronic internal pachymeningitis, is by no means rare. My object has been accomplished if I have succeeded in emphasizing the fact that this condition is worthy of study and not without practical interest.

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[1] Diseases of Infancy, 1897.